PHOTO ROUNDS

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Skin rash and muscle weakness

The patient's facial rash was spreading—and she was having difficulty climbing stairs and brushing her hair

A 48-year-old Hispanic woman came to the clinic as a new patient—her chief complaint was a rash that appeared on her face 3 months before and had recently spread to her chest and hands (**FIGURES 1–3**). It itched occasionally and seemed to worsen after exposure to the sun.

She also said that for the last month she had been feeling very weak—she had difficulty rising from a seated position and walking up the stairs to her apartment. She also felt as if her arms were heavy, making it difficult for her to brush and dry her hair in the morning.

The patient was otherwise healthy with no known medical conditions, and she was not taking any medications. Her family history was noncontributory.

A musculoskeletal examination showed the following:

Upper extremities:

4/5 strength shoulder abduction, internal and external rotation

5/5 strength biceps, triceps, wrist extension/flexion, grip

2+ biceps and triceps deep tendon reflexes bilaterally

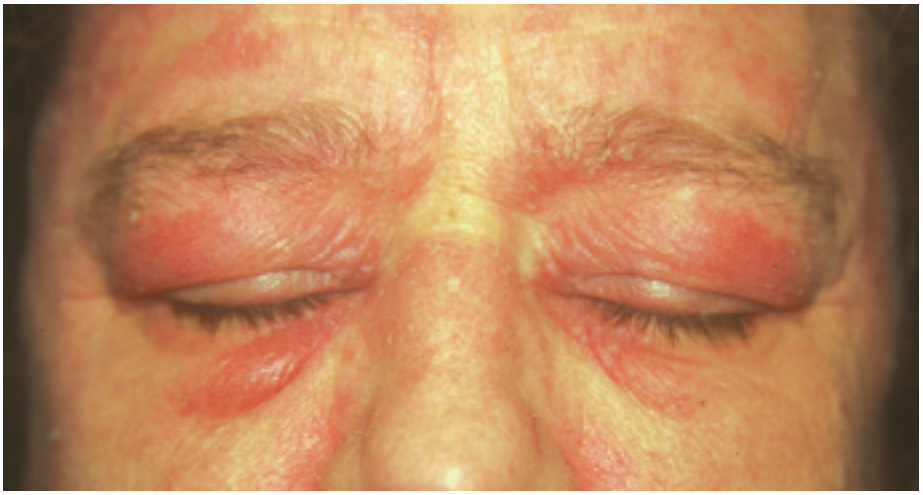
Lower extremities:

4/5 strength hip flexors, quadriceps, hamstrings

5/5 dorsiflexion/plantar flexion

2+ patellar and ankle deep tendon reflexes bilaterally.

FIGURE 1 Facial rash and swelling



Violaceous facial rash with periorbital edema (courtesy of the Division of Dermatology, UTHSCSA).

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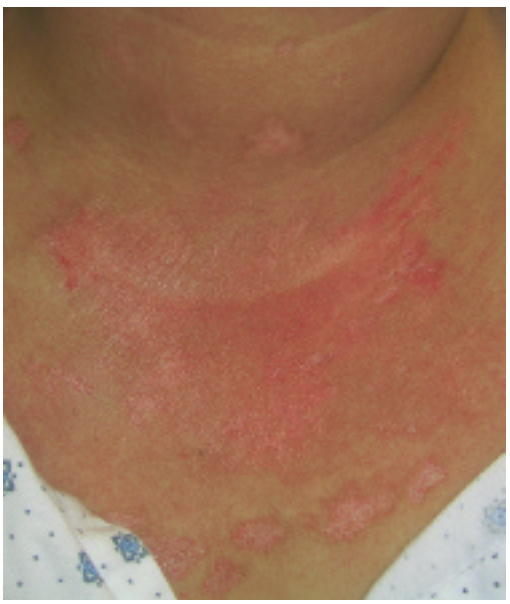
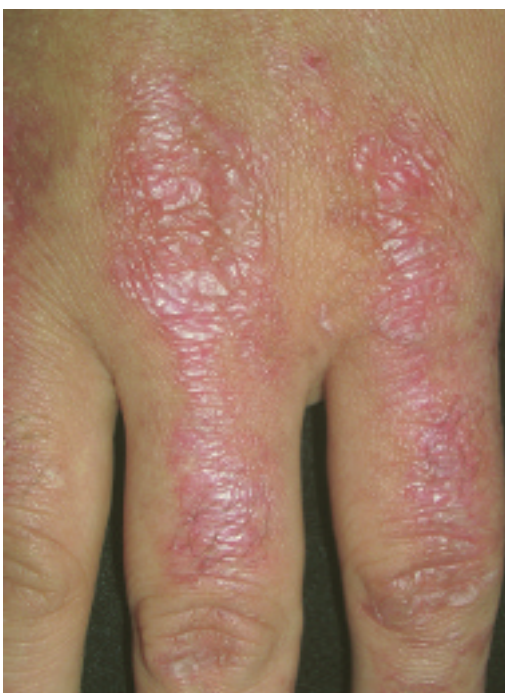
- **What is your diagnosis?**
- **What diagnostic tests would you order for confirmation?**

FIGURE 2 Rash spreading to chest



Mildly pruritic erythematous plaques on the patient's chest.

FIGURE 3 Plaques on knuckles

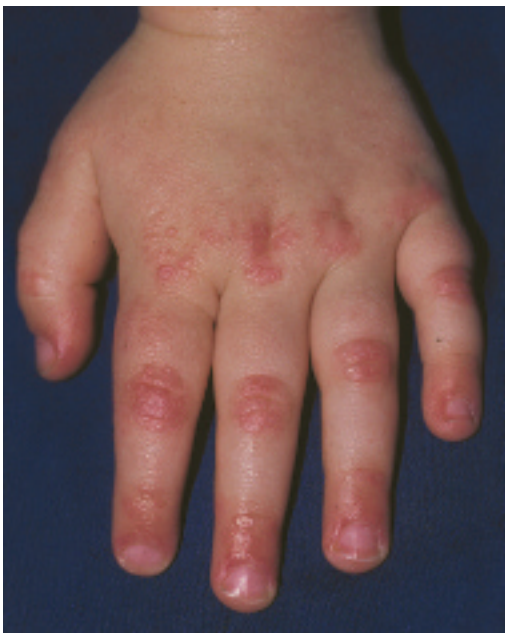


Scaly, erythematous plaques located over the knuckles.

FAST TRACK

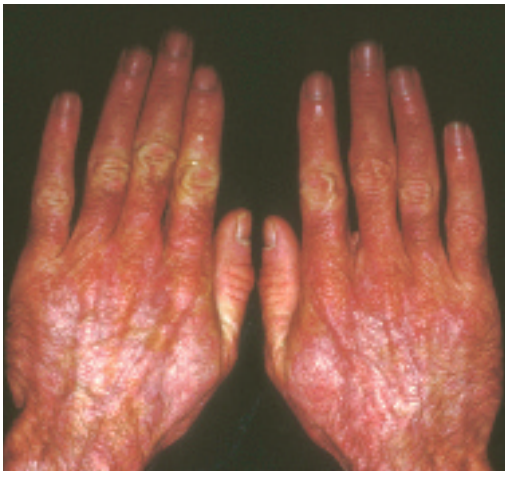
Dermatomyositis patients often complain of difficulty climbing stairs, rising from a seat, or combining their hair

FIGURE 4 Papules on knuckles



Erythematous papules located over the knuckles in a child with dermatomyositis (courtesy of the Division of Dermatology, UTHSCSA).

FIGURE 5 Sparing interphalangeal joints



Erythematous rash sparing the skin overlying the proximal interphalangeal joints in a patient with SLE (Courtesy of the Division of Dermatology, UTHSCSA).

Diagnosis: Dermatomyositis

Dermatomyositis is a systemic disease classified as a type of idiopathic inflammatory myopathy. Dermatomyositis is a rare disease but one that may present initially to the family physician. It affects people at any age but is more commonly seen among children or adults aged >40 years.

Dermatomyositis involves the skin as well as skeletal muscle. Its cause is unknown; however, among those aged >50 years, malignancy may be an underlying cause. Cancers most commonly associated with dermatomyositis include those of the breast, ovary, lung, and gastrointestinal tract.

Skin manifestations may precede, follow, or present simultaneously with muscle involvement. Patients often complain of having difficulty ascending stairs, rising from a seated position, and performing overhead activities such as combing their hair. Patients may or may not have muscle tenderness and atrophy. Patients can have cutaneous involvement for more than a year before developing muscle weakness.[1]

The dermatologic signs of dermatomyositis to watch for:

- *Periorbital heliotrope erythema*, usually associated with edema (**FIGURE 1**).
- *Gottron's papules*—smooth, purple to red papules located over the knuckles, on the sides of the fingers, and sometimes on the elbows and knees. For adults, it is not uncommon to have plaques over the knuckles as opposed to the classic Gottron's papules (**FIGURE 3**). In juvenile-onset dermatomyositis, distinct papules are much more evident upon presentation (**FIGURE 4**). Note that systemic lupus erythematosus (SLE) can present with a rash on the dorsum of the hands, but the rash spares the skin over the metacarpophalangeal and interphalangeal joints and affects the skin between the joints (**FIGURE 5**).
- *Violaceous papular dermatitis with scale*—may occur in localized areas, such as elbows and knees, or be diffusely distributed, starting off as a patchy erythema that coalesces and becomes slightly raised with scale. It tends to be confined to sun-exposed areas and worsens after sun exposure (**FIGURE 2**).
- *Periungual erythema and telangiectasia*—"moth-eaten" cuticles, a characteristic seen in other connective tissue diseases (**FIGURE 6**).[1]

Differential diagnosis

Seborrheic dermatitis—white or yellow, greasy scales on an erythematous base with distribution on scalp, nasolabial folds and chest.

Atopic dermatitis—chronic history of pruritic papules or plaques with scale localized to flexural areas or may be generalized; lichenification may be seen.

Contact dermatitis—papules and vesicles that correspond to contact with allergen.

Polymorphous light eruption—clusters of erythematous, pruritic papules or vesicles occurring most frequently on the neck, anterior chest, arms, and forearms following sun exposure; most common among women in their twenties.

Lichen planus—pruritic, purple, polygonal papules (4 Ps) that may involve hair, nails, and mucous membranes in addition to the skin; more common among women, with onset between 30 to 60 years of age; may last months to years.

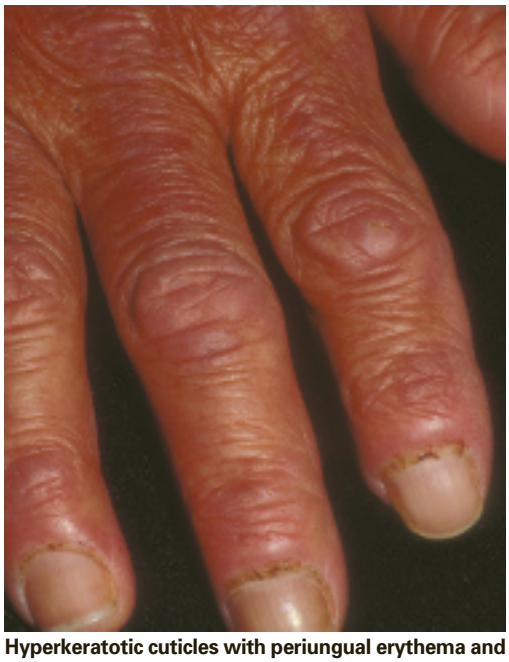
Psoriasis—well-demarcated papules and plaques on an erythematous base with thick, silvery scale; characteristically found on elbows, knees, scalp, nails, and genitalia.

Steroid myopathy—A side effect of systemic steroids, usually seen 4 to 6 weeks after beginning of treatment.

Dermatomyositis-like reaction—onset of similar skin findings with initiation of the following medications and improvement with discontinuation: penicillamine, nonsteroidal anti-inflammatory drugs, and carbamazepine.

Overlap syndrome—The term "overlap" denotes that certain signs are seen in both dermatomyositis and other connective tissue diseases such as scleroderma, rheumatoid arthritis, and lupus erythematosus. Scleroderma and dermatomyositis are the most commonly associated conditions and have been termed sclerodermatomyositis or mixed connective disease. In mixed connective tissue disease, features of SLE, scleroderma, and polymyositis are evident such as malar rash, alopecia, Raynaud's phenomenon, waxy-appearing skin, and proximal muscle weakness.[1,2]

FIGURE 6 Cuticles with erythema



Hyperkeratotic cuticles with periungual erythema and telangiectasia in an adult with dermatomyositis (courtesy of the Division of Dermatology, UTHSCSA).

Diagnostic tests: Muscle enzymes, EMG, biopsy

The diagnosis of dermatomyositis is confirmed by 3 laboratory tests: elevated muscle enzyme levels, electromyography, and muscle biopsy. A punch biopsy is helpful in differentiating dermatomyositis from other papulosquamous diseases such as lichen planus and psoriasis, but be careful as the histology of dermatomyositis is indistinguishable from cutaneous lupus erythematosus.

During the acute active phase, the following serum muscle enzymes may be elevated: creatine kinase (CK), lactate dehydrogenase (LDH), alanine aminotransferase (ALT or SGPT), aspartate aminotransferase (AST or SGOT), and aldolase. CK is elevated among 65% of patients and is most specific for muscle disease.[3] Only one of the aforementioned enzymes may be elevated, so it is necessary to measure them all.

Measuring antibodies such as antinuclear antibody (ANA), Jo-1, SSA (Ro), SSB (La) supports the diagnosis if positive but dermatomyositis cannot be diagnosed sole-

FAST TRACK

Diagnosis: elevated muscle enzymes, evidence of inflammation on EMG, and inflammatory infiltrates on muscle biopsy

ly on positive titers. It is not necessary to obtain an electromyograph or muscle biopsy for a patient with the characteristic skin findings and evidence of elevated muscle enzymes, as the diagnosis of dermatomyositis can be made with confidence. For a patient in whom the presentation is not as straightforward, it may be useful to obtain the electromyograph and muscle biopsy.

Management: Corticosteroids, watch for malignancy

Oral corticosteroids are the treatment of choice (strength of recommendation [SOR]: B).[1,4] Prednisone 0.5 to 1.0 mg/kg body weight per day has been recommended until muscle enzyme levels trend toward normal limits, at which time you can taper the dose. Steroid myopathy is a potential side effect of this treatment regimen; it may occur 4 to 6 weeks after therapy starts.

Several steroid-sparing agents such as methotrexate and cyclosporine are being prescribed by clinicians for dermomyositis but with little published evidence to support effectiveness. Methotrexate is an option for those who do not respond to prednisone or are in need of a steroid-sparing agent secondary to side effects (SOR: C).[2,3] A suggested regimen starts with 7.5 to 10 mg/wk, then increases the dose to 2.5 mg/wk until reaching a total dose of 25 mg/wk. The dose of prednisone should be decreased as the methotrexate dose increases. Azathioprine is another option to consider along with methotrexate, but choosing one agent over another or a combination of 2 agents remains empirical. With any of the immunosuppressants or immunomodulatory agents, it is important to look at their side-effect profiles and monitor the patient accordingly.

After initiating treatment, look for evidence of malignancy among those patients older than 50 years so as not to miss an underlying cancer as the cause for their dermatomyositis. For women without any risk factors, a complete annual physical exam—including pelvic, breast, and rectal exam—is sufficient.[2] It is not necessary to order expensive radiological studies blindly searching for malignancy, especially more than 2 years after the diagnosis is made. The greatest risk of malignancy occurs during the first year after diagnosis with a six-fold increase.[1] The risk drops during the second year and a patient's risk for malignancy is comparable to the normal population in the years following. A mammogram and colonoscopy might be indicated after considering the patient's age and family history.

The patient's treatment and outcome

The patient was started on 60 mg of oral prednisone, taken in a single daily dose. She also began physical therapy twice a week in order to prevent muscle atrophy and maximize function. She took the prednisone for 1 month, at which time her creatine kinase level was trending towards normal. We then began slowly tapering the prednisone over the next 6 months.

She reported improvement in her strength 3 months after starting the systemic steroids. Little improvement was seen in the patient's skin while on systemic steroids, but after prescribing 0.1% triamcinolone ointment, recommending a broad-spectrum sunscreen, and limiting sun exposure, the patient reported less erythema and edema. ■

FAST TRACK

After start of treatment, look for evidence of malignancy in patients older than 50 years

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